Case Reports

Recurrent Asystole in a Child

HERBERT A. LASSITER, MD
Riverton, Wyoming

SEIZURES DUE to recurrent cardiac syncope in an otherwise healthy child are rare. In this report I describe the case of a 10-year-old boy with a structurally normal heart who had seizures due to recurrent sinoatrial pauses* with asystole.

Report of a Case

The patient presented to Riverton (Wyoming) Memorial Hospital having "passed out." He had been leaning against a well pump when he fell to the ground. His arms and legs jerked for five seconds and he was incontinent of urine and feces. He became alert immediately. Two weeks previously while camping, he had awakened, walked from his tent, lost consciousness while walking and had a seizure identical to that described above. He said he had had no premonitory symptoms or aura.

During the previous two months, he reported two episodes of rapid heart rate associated with chest pain lasting five minutes each. He did not seek medical attention for the first seizure described above, nor for the episodes of tachycardia.

He had previously been in excellent health. There was no family history of cardiac disease. A maternal aunt has recurrent seizures due to a stroke at age 19. At 8 years of age the patient had a left herniorrhaphy without complications. He had not been taking any medications in recent months.

On physicial examination he appeared thin and was pleasant, alert and cooperative. His temperature was 37.2°C (99.0°F), pulse rate 96 per minute, respirations 20 and blood pressure 96/60 mm of mercury. Supine and standing blood pressures were identical. There was a well-healed left herniorrhaphy scar. The scrotal sac was empty on the right. On cardiac examination he had normal heart sounds and no murmur. The remainder of the physical examination, including the neurologic examination, elicited no abnormalities.

*The term "sinoatrial pause" is preferred to "sinus arrest" because sinus arrest cannot be distinguished from sinoatrial block by standard electrocardiography.

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Dr Lassiter is in private practice in Riverton, Wyoming.
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Reprint requests to Herbert A. Lassiter, MD, 511 North 12th East, Riverton, WY 82501.

After admission to hospital, an irregular pulse rate of 52 per minute was noted. An electrocardiogram (ECG) and rhythm strip showed sinus arrhythmia, periods of sinus bradycardia and a wandering atrial pacemaker. The ECG findings were otherwise normal (Figure 1). Telemetry showed heart rates as low as 38 per minute during sleep, with PR intervals varying from 0.10 to 0.18 seconds with inconsistency in the configuration of the P wave.

Chest and skull x-ray films and electroencephalogram (EEG) showed normal findings. Laboratory studies gave the following values: glucose, 75, calcium, 9.5, and uric acid, 6.0 mg per dl; total protein, 7.3, albumin, 4.1, and globulin, 3.2 grams per dl; cholesterol, 174, blood urea nitrogen, 15, creatinine, 0.7, and total bilirubin, 0.6 mg per dl; alkaline phosphatase, 153, alanine aminotransferase (ALT, formerly glutamic-pyruvic transaminase), 7, aspartate aminotransferase (AST, formerly glutamic-oxaloacetic transaminase), 7, lactic dehydrogenase, 80, and creatine phosphokinase, 84 IU per liter; sodium, 143, potassium, 4.2, chloride, 109, and carbon dioxide, 24 mEq per liter; leukocyte count, 6,900 per μl with a normal differential count; hemoglobin, 13.5 grams per dl; hematocrit, 38.9%; erythrocyte sedimentation rate, 6 mm an hour, and platelet count, 241,000 per μl.

At 5 AM on the third day of his hospital stay, while being sleep-deprived for a second EEG, telemetry recorded profound bradycardia followed by prolonged asystole (Figure 2). After the development of asystole, the child was found in his chair having a generalized tonic seizure that ceased spontaneously within a few seconds. Shortly thereafter a temporary pacemaker was implanted. During the cutaneous infiltration of lidocaine in preparing to insert the pacemaker with the patient in the supine position, he had asystole for 20 seconds during which he again suffered a brief tonic seizure, terminated by the return of sinus rhythm.

Subsequently, cardiac evaluation of the patient was done at the Mayo Clinic in Rochester, Minnesota, including electrophysiologic studies, which showed normal sinoatrial node function, normal intrinsic heart rate (heart rate after autonomic blockade with atropine sulfate and propranolol hydrochloride), accelerated atrioventricular nodal conduction, dual atrioventricular nodal pathways without inducible reentry tachycardia and inducible sustained atrial flutter terminated by atrial pacing. Table 1 contains a summary of the sinoatrial nodal function. Neurologic and genetic consultants found no evidence of neurologic disease or an identifiable syndrome. Serum triglyceride values were elevated at 214 mg per dl. The serum thyroxine level

ABBREVIATIONS USED IN TEXT

ECG = electrocardiogram
EEG = electroencephalogram

was normal at 8.3 mg per dl. He was discharged from the Mayo Clinic on a regimen of propantheline bromide, 7.5 mg four times a day, and a low-fat low-carbohydrate diet.

Ten days later, a few seconds after blowing a flute at home, he had another syncopal attack despite the medication. A permanent demand ventricular pacemaker was implanted at Primary Children's Hospital, Salt Lake City, with the rate set at 50. Three months after pacemaker implantation he underwent right orchiopexy without complication.

Six months later, at school, the patient had 20 minutes of rapid heart rate associated with pallor, weakness and dizziness. He was admitted for 48 hours of inpatient observation and monitoring, which showed continued sinus arrhythmia, sinus bradycardia and normal pacemaker function, but tachycardia was not documented. Seven months after implantation, his demand rate was reprogrammed at 66 to alleviate recurrent symptoms of dizziness. He has suffered neither seizures nor syncope since implantation of his pacemaker.

Discussion

The differential diagnosis of generalized motor seizures must include cardiac syncope due to dysrhythmias even in children. Although sinus bradycardia and sinus arrhythmia are common in children, these findings must not be dismissed out of hand when faced with a history of cerebral dysfunction such as dizziness, fainting, memory loss or seizures.

This patient manifested many of the clinical signs and symptoms of the sick sinus syndrome—sinus bradycardia, sinoatrial pauses, syncope, seizures and a history suggestive of tachydysrhythmias. However, results of his electrophysiologic studies ruled out sick sinus syndrome due to intrinsic dysfunction of the sinus node by virtue of his normal response to atropine therapy with a brisk sinus tachycardia and his normal corrected sinus node recovery time. The sinus node recovery time is measured by subjecting the heart to rapid atrial pacing (starting at 120 beats per minute and increasing the rate by increments to 200) and measuring the longest interval from the last atrial pacemaker-induced spike to the onset of the first P wave (the "PP" interval). The sinus node recovery time exceeds the baseline PP in-

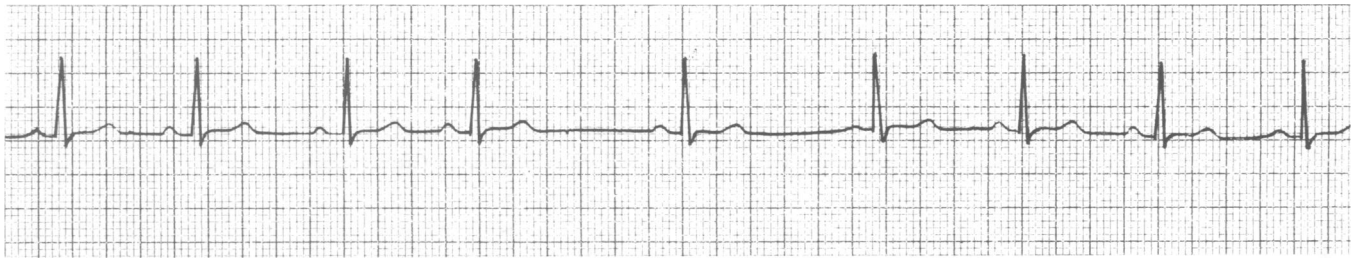

Figure 1.—Lead II rhythm strip shows sinus arrhythmia and varying configuration of the "P" wave (wandering atrial pacemaker). The QRS has been visually enhanced.

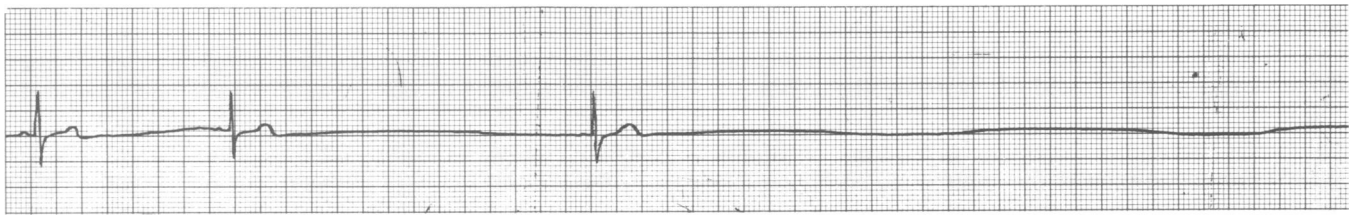

Figure 2.—Rhythm strip via telemetry shows severe sinus bradycardia and sinus arrest. The QRS has been visually enhanced.

TABLE 1.—*Summary of Sinus Node Function*

	Autonomic Blockade		
	Pre-Atropine Sulfate	*Post-Atropine Sulfate*	*Post-Atropine and Propranolol Hydrochloride*
Resting heart rate (beats/min)	88	138 (normal, 114)*	118
Longest sinus node recovery time† (msec)	970 (normal, 1,130)*		680
Longest corrected sinus node recovery (msec)	290 (normal, 525)‡		175

*Normal values taken from Yabek.[3]
†Atrial pacing ranged from 120 to 200 beats per minute.
‡The Mayo Clinic sets the upper limit of normal for the corrected sinus node recovery time at 525 msec.

terval in normal hearts, but is excessively prolonged when the sinus node is diseased or damaged. A prolonged sinus node recovery time strongly suggests decreased automaticity of the pacemaker cells in the sinus node or a delay in the exit of electrical impulses from the node into the atria due to disease, usually fibrosis, in the perinodal tissue. The computation of the sinus node recovery time—that is, recovery time minus the baseline PP interval—corrects for the fact that the sinus node recovery time is directly related to the baseline PP interval (the slower the resting heart rate, the longer the recovery time).

The normal sinus node recovery time, corrected sinus node recovery time and response to atropine effectively rule out an intrinsically dysfunctional sinus node as an explanation for the patient's findings.[1-9]

Vasodepressor, or vasovagal, syncope is a common phenomenon, but does not adequately account for this patient's symptoms. Vasodepressor syncope typically occurs in situations that a patient finds distasteful, is characterized by a presyncopal phase in which the patient anticipates losing consciousness and occurs in the standing or sitting position, only rarely in the recumbent position. Peripheral pooling of venous blood is the primary mechanism leading to hypotension. Vagal-induced bradycardia contributes to the decreased cerebral perfusion, but sinoatrial pause and asystole are not part of the syndrome.[10] Vagovagal syncope occurs when cardiac asystole occurs as a result of a reflex that is located entirely within the vagus nerve. However, this patient has no known lesion that might account for this phenomenon such as an esophageal diverticulum prone to distention, glossopharyngeal neuralgia, cardiospasm or a lesion in the larynx, pleura or peritoneum.[10]

In short, the patient in the case reported here does not seem to fit the description of patients previously classified as having one of the variants of cardiac syncope. Although it is doubtful that the dysrhythmia described is unique, an extensive search of the literature failed to reveal a case of recurrent sinoatrial pauses, asystole and seizures in an otherwise healthy child with a structurally normal heart and a normally functioning sinus node when tested electrophysiologically.

Autonomic imbalance appears to be at the root of this child's dilemma. Excessive vagal tone or diminished sympathetic tone at the sinus node could explain the findings as could excessive cholinergic sensitivity or diminished adrenergic sensitivity of his sinus node pacemaker cells. Aside from the fact that autonomic control of his sinus node could be blocked by administering atropine and propranolol, little is known of his intrinsic cholinergic control mechanisms.

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Peutz-Jeghers Syndrome With Cervical Adenocarcinoma and Enteritis Cystica Profunda

NOLAN J. ANDERSON, MD
ERNESTO S. RIVERA, MD
DONALD J. FLORES, MD
Delano, California

THE PEUTZ-JEGHERS SYNDROME, one of the familial polyposes syndromes, is an autosomal dominant condition characterized by mucocutaneous melanin pigmentation and gastrointestinal polyps.[1,2] It is now recognized that there is a high risk that a distinctive ovarian tumor described as a sex cord tumor with annular tubules may develop in cases of this disorder.[3,4] Lately, however, a rare, well-differentiated adenocarcinoma of the cervix with a very poor five-year survival rate has also been reported with this condition.[5,6] An equally rare intestinal lesion known as enteritis cystica profunda is occasionally found in patients with Peutz-Jeghers syndrome.[7] This latter lesion is characterized by mucosal glands and mucinous cysts in the small and large bowel that penetrate the tunica muscularis. It sometimes involves the entire wall up to the serosa and, thus, has been mistaken for adenocarcinoma. We report the first case of Peutz-Jeghers syndrome that is associated with cervical adenocarcinoma and enteritis cystica profunda.

Report of a Case

The patient, a 29-year-old Latin-American woman, was admitted for weakness, dizziness and lower abdominal pain for one week. She gave a history of irregular and prolonged menstruation since the age of 16. Three months before admission, she had a spontaneous abortion of a 10-week-old pregnancy for which she required transfusion of two units of blood. She later had daily vaginal bleeding with lower abdominal pain for one month, culminating in this admission.

She was very pale, with a hemoglobin of 5.8 grams per dl and hematocrit of 17%. Pigmentation about the

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From the Departments of Surgery (Dr Anderson), Pathology (Dr Rivera) and Obstetrics and Gynecology (Dr Flores), Delano Regional Medical Center, Delano, California.

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Reprint requests to Ernesto S. Rivera, MD, Dept of Pathology, Delano Regional Medical Center, PO Box 460, Delano, CA 93216.